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## Case report

## Unilateral pulmonary agenesis presenting in adulthood

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Chronic respiratory symptoms with unilateral opaque hemithorax should include agenesis of lung in its differential diagnosis

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## ABSTRACT

Agenesis of lung, a rare congenital anomaly, may present in adult life with features of recurrent chest infections and radiologically may mimic many common conditions presenting as opaque hemithorax with ipsilateral shifting of mediastinum. Here, a case of a young man presenting with frequent attacks of cough expectoration and progressive dyspnoea since childhood, proved to be a case of left pulmonary agenesis on CT scan and bronchoscopy, is to be discussed.

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## 1. Introduction

The term 'agenesis' is taken to mean Partial or almost complete absence of growth in the lung.<sup>1</sup> The rarity of this condition is evident by the infrequent reporting of such cases in literature with prevalence of 34 per million live births. Till 1970 only 220 cases were reported world wide. Needless to say, bilateral agenesis is incompatible with life. Unilateral agenesis of the lung is much less rare and may present with varying degrees of severity. They are often wrongly diagnosed for more common conditions of unilateral volume loss and it is even more challenging if it comes to notice in adult life. Here we report a case of young man presenting with left pulmonary agenesis.

## 2. Case history

A 24 year old male presented with insidious onset, progressive shortness of breath since childhood and frequent episodes of cough with mucopurulent sputum, often one cupful per day, yellowish in colour. There were no history of orthopnea, palpitation, wheezing, chest pain, coughing out of blood, anorexia and weight loss. He had no past history suggestive of pulmonary tuberculosis. His perinatal history was insignificant and no history of similar complaints in any

of his siblings. On examination, he was an average built male, malnourished, preferring left lateral decubitus. Pallor, icterus, clubbing, engorged neck veins and lymphadenopathy were absent. Central cyanosis and pitting pedal oedema were present. On inspection of chest, accessory muscles were working, drooping of shoulder seen in left side and scoliosis with convexity to right noticed. Intercostal suction was seen. On palpation, movement diminished in left side with rib crowding, trachea deviated to left and apex beat placed at left 6th intercostal space in mid axillary line. Expansion of chest was 3 cm and vocal fremitus diminished throughout the left side. On percussion, left side had impaired note 7th ICS downward along MAL and scapular line, resonant in rest of the areas. On auscultation, bilateral vesicular breath sound heard with reduced vocal resonance on left side, bilateral coarse crepitations heard in inter and infrascapular area and right axillary region. Liver was palpable by 2 cm. S<sub>2</sub> was loud, other systems were within normal limits.

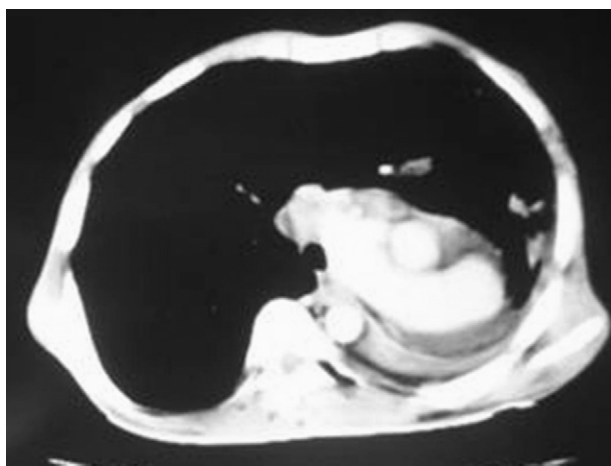
Chest radiograph showed homogenous opacity in the left lower zone, obliterating the left costophrenic angle with gross shifting of the mediastinum to the left and scoliosis with convexity to the right and reticulonodular shadows in the right lower zone (Fig. 1). ECG showed tall peaked P waves in lead 2. Echodoppler study revealed moderate pulmonary hypertension (65 mm Hg), right ventricular hypertrophy and mild pericardial effusion. Contrast enhanced computed tomogram showed absence of left lung and herniation of right lung to the left and bronchiectatic changes in right lung (Fig. 3), absence of left main bronchus and left pulmonary artery (Fig. 2). Fibreoptic bronchoscopy showed only a dimple at the place of the opening of left main bronchus (Fig. 4). Ultrasonogram revealed pericardial effusion. He was diagnosed as left sided

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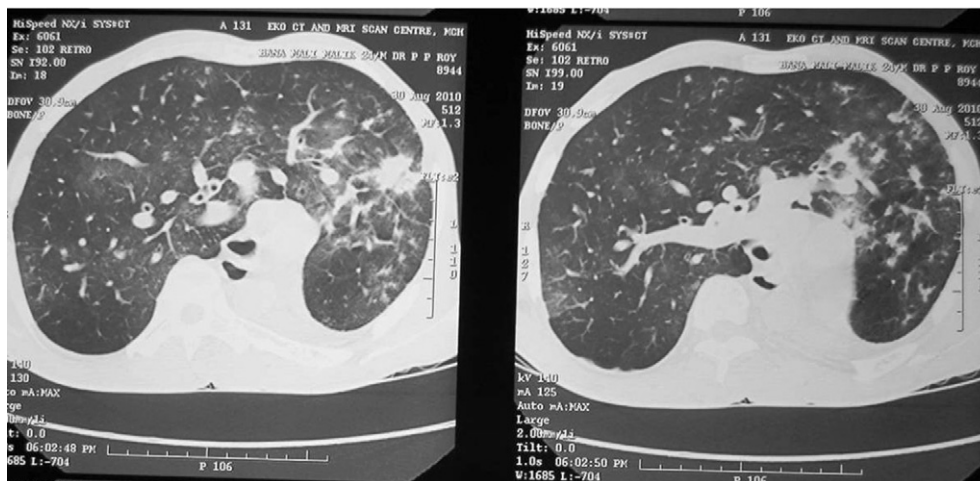
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**Fig. 1.** Chest Radiograph showing homogenous opacity in the left lower zone,obliterating the left costophrenic angle with gross shifting of the mediastinum to the left and scoliosis with convexity to the right and reticulonodular shadows in the right lower zone.



**Fig. 2.** Mediastinal window with contrast showing absence of opening of left main bronchus and left pulmonary artery not seen.



**Fig. 3.** Parenchymal window of contrast enhanced CT scan chest showing absence of left lung and herniation of right lung to the left,also there are bronchiectatic changes.

agenesis of lung with left pulmonary artery atresia, pulmonary artery hypertension and bronchiectasis of right lung.

### 3. Discussion

Unilateral agenesis of the lung may be present to varying degrees of severity. The left lung is affected more frequently than the right, males predominate over females and the majority of cases exhibit other congenital abnormalities like patent ductus arteriosus, pulmonary artery atresia, cardiac malformation, tracheo-esophageal fistula, cardiac malformation and horse-shoe kidney. However, several older reports prove that other anomalies are more associated with right sided agenesis and persons with right sided agenesis mostly die within first year of their life, due to associated cardiac malformations.<sup>2</sup> Originally *Schneider (1912)*<sup>3</sup> classified agenesis into three groups which was later on modified by *Boyd*<sup>4</sup> as-

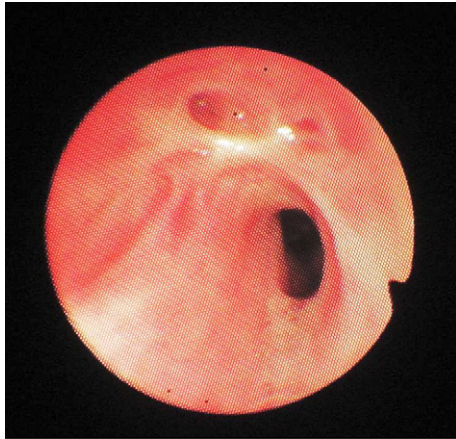
Type I (Agenesis): Complete absence of lung and bronchus and absence of blood vessels to the affected side.

Type II (Aplasia): Rudimentary bronchus with complete absence of lung tissue.

Type III (Hypoplasia): Presence of variable amounts of lung parenchyma, bronchial tree and supporting vasculature.

Our patient has been classified as Type I. In *Schneider's* agenesis grade I and II, the affected side contains no lung tissue, and only the existing lung gets the branch from the main pulmonary artery, an observation which has been confirmed several times and has been seen in our case also. Clinical presentation of agenesis lung is marked by its variety from recurrent childhood respiratory infection resulting from imperfect drainage of lung secretions or from the spillover of pooled secretions from a blind bronchial stump into initially normal lung tissue, frequent haemoptysis due to bronchiectasis of remaining lung to major organ malformation leading the patient to succumb in early life. A similar case was reported in Turkey as, a 30-year-old man presenting with dyspnoea was diagnosed to have right lung agenesis and left pulmonary bronchiectasis.<sup>5</sup>

Autosomal recessive chromosomal aberration, associated with consanguineous marriage<sup>6</sup>, deficiency of vitamin A, intrauterine infections, environmental factors have been held responsible for the etiology of congenital lung malformations. During normal development, the heart shifts to the left in the 4th week of foetal life and simultaneously the trachea develops as a ventral diverticulum arising from the foregut.<sup>7</sup> Pulmonary agenesis or aplasia occurs perhaps due to the failure of the bronchial analogue to



**Fig. 4.** Dimple at the place of opening of left main bronchus.

divide equally between the two lung buds. If this balance is not established, one side will develop normally while the other will fail completely (agenesis/aplasia) or undergo only limited development (dysplasia or hypoplasia).

In adults, unilateral agenesis of lung may mimic collapse, thickening of pleura, destroyed lung, pneumonectomy, scoliosis with pleural effusion, diaphragmatic hernia, adenomatoid cystic malformations and sequestrations.

CT Chest, which provides detailed description of bronchial tree, parenchyma and vasculature is considered to be the most definitive investigation to diagnose agenesis when chest radiograph is not diagnostic.<sup>8</sup> Bronchography is almost obsolete now, but

bronchoscopy is useful to demonstrate rudimentary bronchus. Pulmonary angiography or MRI Angiography is considered to show the absence of ipsilateral pulmonary vessel and cardiac catheterization may be needed to rule out cardiac malformations and to quantify Pulmonary artery pressure. In our case these could not be done as the patient could not afford them.

No treatment is required in asymptomatic cases. Treatment is necessary for recurrent chest infections. Patients having bronchial stumps may require surgical removal if postural drainage and antibiotics fail to resolve the infection. Corrective surgery of associated congenital anomalies, wherever feasible, may be undertaken.<sup>9</sup>

#### Conflict of interest statement

We have no conflict of interest regarding the article.

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